

LETTERS TO THE EDITOR

New variant Creutzfeldt-Jakob disease presenting with loss of taste and smell

Abnormalities of smell and taste have been described in some neurodegenerative diseases including Alzheimer's dementia, idiopathic Parkinson's disease, Huntington's chorea, Korsakoff's syndrome, Pick's disease, the parkinsonian dementia complex of Guam, and amyotrophic lateral sclerosis.¹ Hyposmia and hypogeusia are a feature of normal aging but they have not been recorded as a prominent early feature in previous reports of variant Creutzfeldt-Jakob disease (vCJD).²⁻⁵ We describe a patient with vCJD whose first symptoms included deficits of taste and smell.

At the time of his initial neurological assessment, this 54 year old ceramic tiler had a 12 month history of loss of taste and smell, anxiety, low mood, and unusually short temper. He first became aware that something was wrong when he lost the ability to differentiate the taste of tea from that of beer. Loss of taste and personality change progressed gradually. He began to crave vanilla ice cream although he had never liked sweet foods in the past. He became fearful of leaving his house. Six months into his illness he became increasingly sleepy. On average he would sleep for 12 hours a day although he could sleep for 20 hours at a time. He developed slurred speech, unsteady gait, upper limb tremor, and impotence. Neither the patient nor his wife noticed any change in his memory.

On examination the patient had scanning dysarthria. His abbreviated mini mental test score was 7/10 (unable to remember an address, give his age, or name the year). He had a staring look with limitation of upgaze. Limb tone was increased with brisk tendon reflexes, ankle clonus, and bilaterally extensor plantar responses. There was mild upper limb ataxia and severe ataxia of gait. Although spinothalamic sensation was intact, joint position sense was impaired in both lower limbs. Olfactory testing disclosed evidence of impaired smell detection and recognition. The patient thought that he could smell something when tested with lavender and tar essence but was unable to recognise or describe the smell.

Over the next 4 months the patient deteriorated rapidly with progressive ataxia, confusion, and agitation. He developed myoclonic jerks 2 weeks before dying of bronchopneumonia 16 months after the onset of his first symptoms.

Our investigations in vivo supported a diagnosis of vCJD (suggestion of high signal within the thalamus on T2 weighted cranial MRI, negative immunoassay for 14-3-3 protein but raised S100b protein in the CSF (0.91 ng/ml, reference range <0.38 ng/ml), no known mutations in the prion protein gene, methionine homozygous at codon 129). Differential diagnoses were excluded by normal or negative full blood count, erythrocyte sedimentation rate, B12, folate, thyroid function tests, treponemal serology, antinuclear factors, glucose, antineuronal, antiPurkinje

cell, antithyroid, antigliadin, and anti-endothelial antibodies, serum electrophoresis, immunoglobulins, bone marrow aspirate, and trephine. Several EEGs showed diffuse slowing of background rhythms.

Postmortem neuropathological and histological examination confirmed the diagnosis of vCJD. In addition, there was evidence of bronchopneumonia. Sections of brain (brain weight 1322 g) showed extensive tissue involvement with prominent neuronal loss, astrocytosis, spongiform change, and numerous Kuru-type plaques, including florid plaques. These changes were seen in the cortices of frontal, parietal, temporal, and occipital lobes, basal ganglia, thalami, periventricular grey matter, brain stem, olfactory areas of the cerebrum, and the cerebellar cortex. Spongiform change and plaques were most prominent in the cortical regions and cerebellum. Plaques were particularly dense in the molecular and granular layers of the cerebellar cortex. Neuronal loss, gliosis, and spongiform change were most conspicuous in

the basal ganglia and thalami. Immunohistochemistry for prion protein (PrP KG9 1:150, monoclonal antibody, courtesy of the CJD surveillance centre in Edinburgh) showed prominent staining in the plaques and diffusely in the neuropil of cerebral and cerebellar cortices. The olfactory tract showed prominent and diffuse staining for prion protein (PrP) associated with vacuolation (fig 1).

It is difficult to determine the precise cause of our patient's olfactory and gustatory dysfunction. There were significant histopathological changes in the basal forebrain where both taste and smell are represented.² Deposits of prion were, however, also found in many other parts of the brain involved in the neural processing of these senses. Changes were particularly prominent in the olfactory tract. Our patient illustrates that our understanding of the clinical range of vCJD remains incomplete. Loss of taste and smell are not at all specific to vCJD. These symptoms can have many causes including

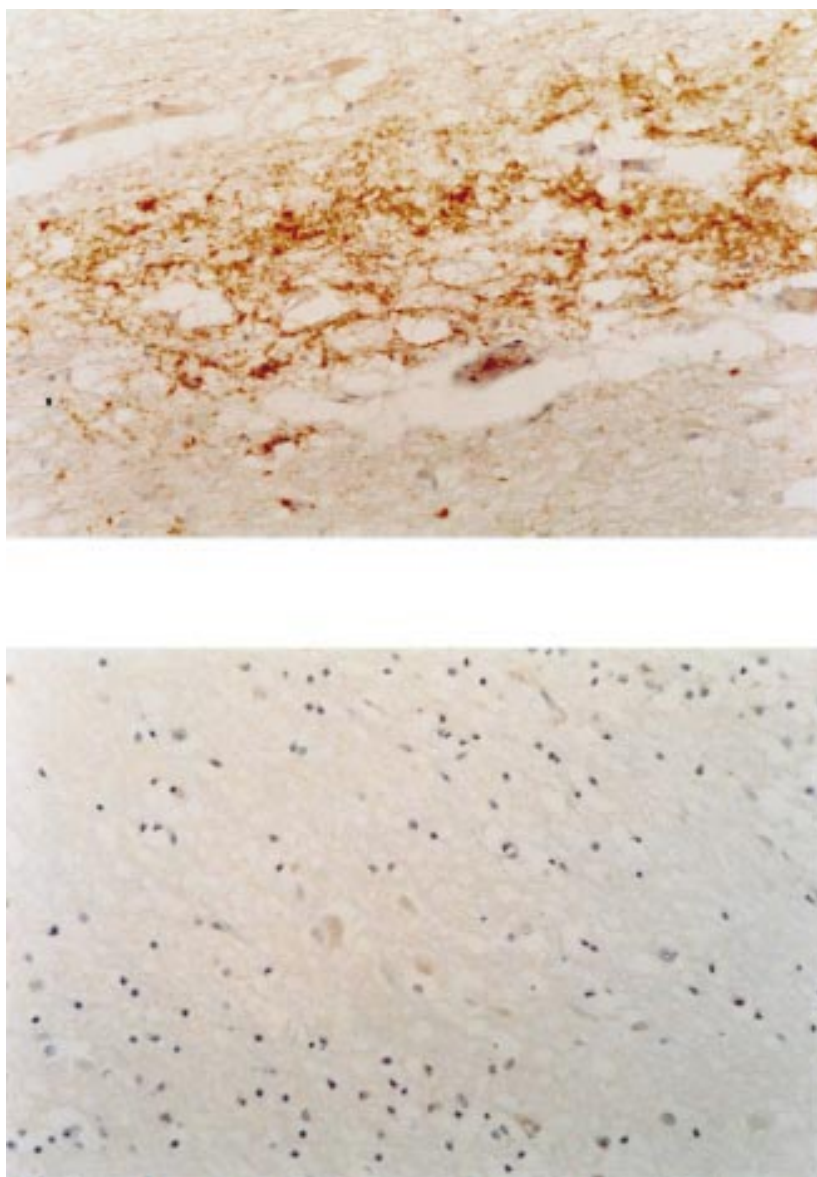


Figure 1 Bottom: normal (control) olfactory tract of a 61 year old man showing no prion protein (PrP) deposition after immunostaining with PrP. Top: olfactory tract showing vacuolation and prominent diffuse staining for PrP. PrP plaques and spongiform changes were also found in other parts of the olfactory system, the cortex, basal ganglia, cerebellum, and brain stem.

other neurodegenerative disorders. However, the diagnosis of vCJD should be considered if hypogeusia and hyposmia are accompanied by changes of personality and other, more "typical" features of vCJD.

M REUBER

A S N AL-DIN

Department of Neurology, Pinderfields General Hospital, Aberford Road, Wakefield WF1 4DG, UK

A BABORIE

A CHAKRABARTY

Department of Neuropathology, General Infirmary at Leeds, Great George Street, Leeds LS1 3EX, UK

Correspondence to: Dr A S N Al-Din

- 1 Meshulam RI, Moberg PJ, Mahr RN, *et al.* Olfaction in neurodegenerative disease: a meta-analysis of olfactory functioning in Alzheimer's and Parkinson's diseases. *Arch Neurol* 1998;55:84-90.
- 2 Schiffman SS. Taste and smell losses in normal aging and disease. *JAMA* 1997;278:1357-62.
- 3 Will RG, Zeidler M, Stewart GE, *et al.* Diagnosis of new variant Creutzfeldt-Jakob disease. *Ann Neurol* 2000;47:575-82.
- 4 Allroggen H, Dennis G, Abbott RJ, *et al.* New variant Creutzfeldt-Jakob disease: three case reports from Leicestershire. *J Neurol Neurosurg Psychiatry* 2000;68:375-8.
- 5 Zeidler M, Stewart GE, Barraclough CR, *et al.* New variant Creutzfeldt-Jakob disease: neurological features and diagnostic tests. *Lancet* 1997;350:903-7.

Obsessive-compulsive characteristics in patients with writer's cramp

Writer's cramp is characterised by a muscular spasm in the hand of the writing arm, and is often provoked during specific tasks such as writing. Because of its highly task specific nature, some neurologists believed that writer's cramp was of psychogenic origin. However, recent electrophysiological and neuroimaging studies unanimously confirmed basal ganglia dysfunction in patients with writer's cramp. As a consequence, writer's cramp is currently regarded as a form of focal dystonia with neurophysiological pathogenetic mechanisms.¹

On the other hand, in the classic psychiatric literature, clinicians sporadically pointed out obsessive-compulsive personalities in patients with writer's cramp. Bindman and Tibbetts described 10 patients with writer's cramp, nine of whom had obsessional personalities.² These findings have never been confirmed using reliable psychometric measures. However, because of the above mentioned evidence of basal ganglia pathophysiology in writer's cramp, together with the growing evidence of basal ganglia involvement in obsessive-compulsive disorder, it is of great interest to elucidate the relation between writer's cramp and obsessive-compulsive symptoms.

In the present study, we evaluated obsessive-compulsive symptoms in patients with writer's cramp. Obsessive-compulsive symptoms may simply be a psychological reaction to the writing impairment. To rule out this possibility, patients with writing impairment due to peripheral nerve lesions were included as a second group of control

subjects, in addition to normal healthy control subjects.

Twelve consecutive patients with writer's cramp who visited the dystonia outpatient clinic of the Department of Neurology at Kyoto University from May 1998 to December 1998, were evaluated with the Yale-Brown obsessive-compulsive scale (Y-BOCS). All patients had been treated with muscle afferent block for more than 3 months.³ As a consequence, their writing disabilities were moderately improved. All of them could write their names in four to six Chinese characters at the time of assessment. A disease control group consisted of seven patients with carpal tunnel syndrome, three with cervical spondylotic radiculopathy, one with chronic inflammatory demyelinating polyneuropathy, and one with myasthenia gravis. All these patients showed disabilities in finger movements due to peripheral nerve lesions of an upper limb, and were recruited from the outpatient clinic of the Department of Neurology at Kyoto University. A healthy control group, age and sex matched to the writer's cramp group, consisted of office clerks or secretaries of Kyoto University and Shiga University. None of the subjects had received antidepressive or neuroleptic medication for at least 6 months before the study. None of the subjects had a history of any psychiatric diseases.

For these three groups, obsessive-compulsive symptoms were evaluated by trained psychiatrists, based on the semistructured interviews assessing Y-BOCS. The scores of the writer's cramp group and two control groups on Y-BOCS were compared using a two tailed Kruskal-Wallis test for non-parametric data. p Values lower than 0.05 were considered to be significant. When the result was significant, the analysis was repeated with each pair of groups, and the p values were multiplied by three to correct for multiple comparisons (two tailed Mann-Whitney tests). Informed consent was obtained from all the participants after the procedure of the study had been fully explained.

The 12 patients with writer's cramp, 12 patients in the disease control group, and 12 normal controls did not differ in age or sex ratio (table 1). Mean Y-BOCS scores were 5.8 (SD 5.5, range 0-18) for the writer's cramp group, 1.0 (SD 1.4, range 0-4) for the disease control group, and 1.0 (SD 1.0, range=0-2) for the normal control group, respectively. There was a significant effect of diagnosis on the total scores of Y-BOCS (Kruskal-Wallis test, $\chi^2=5.99$, $p=0.012$), with significant differences between the writer's cramp group and disease control group ($p=0.030$, corrected for three comparisons) or normal control group ($p=0.036$) but not between the disease control group and the normal control group ($p>0.99$). The compulsive symptoms of the writer's cramp group were mainly related to cleaning and ordering.

The result of our study showed that patients with writer's cramp had higher obsessive-compulsive symptoms than the disease control group or normal control group.

Although according to the classic literature the patients with writer's cramp often have obsessive personality features,² the present study is the first to confirm the clinical finding using objective psychometric measures.

The disease control group consisted of patients with peripheral nerve lesions, who had discomfort in writing comparable with the patients with writer's cramp in this study, who were treated with muscle afferent block.³ Thus the current result does not support the idea that physical distress and social disability due to writing impairment is the main cause of the obsessive-compulsive features of patients with writer's cramp, but supports the notion that writer's cramp and obsessive-compulsive symptoms would develop due to common pathophysiological mechanisms. However, it should be noted that our patients with writer's cramp had been under the treatment for more than 3 months and the duration of disease was relatively short. In most cases of writer's cramp without treatment, it is known that writing disability is extremely severe, causing a significant effect on their lifestyle. The present result does not deny the possibility that the condition could cause abnormal psychopathology in writer's cramp.

Recent studies reported abnormal metabolic activity in basal ganglia together with frontal and anterior cingulate areas in patients with obsessive-compulsive disorder. These data suggest basal ganglia dysfunction as a neural basis of obsessive-compulsive symptoms (for a review, see Saxena *et al*).⁴ Therefore, we suspect that basal ganglia dysfunction is the common neural basis of writer's cramp and obsessive-compulsive symptoms. In other forms of focal dystonia such as spasmodic torticollis and blepharospasm, high obsessive-compulsive symptoms were reported.⁵ Thus, the basal ganglia hypothesis of obsessive-compulsive symptoms may be applied to focal dystonia in general. As obsessive-compulsive symptoms of the patients in this study were not severe enough for the diagnosis of obsessive-compulsive disorder, and due to the small sample size, these findings should be interpreted with caution. Larger studies are necessary in clarifying the pathophysiology of writer's cramp and associated obsessive-compulsive symptoms.

Y KUBOTA

T MURAI

T OKADA

A HAYASHI

Department of Neuropsychiatry, Faculty of Medicine, Kyoto University, 54 Shogoin-Kawaharacho, Kyoto 606, Japan

M TOICHI

Division of Child and Adolescent Psychiatry, Case Western Reserve University, University Hospitals of Cleveland, Cleveland, USA

M SAKIHAMA

Minakuchi Hospital, Shiga, Japan

T SAKAMOTO

K ASANUMA

S MATSUMOTO

R KAJI

Department of Neurology, Faculty of Medicine, Kyoto University, Kyoto, Japan

Correspondence to: Dr Y Kubota

Table 1 Ages, disease duration, and sex ratio of writer's cramp group and control groups

	Age range (y)	Mean age (SD) (y)	Disease duration range (y)	Mean duration (SD) (y)	Sex ratio (male/female)
Writer's cramp	21-70	42.6 (16.5)	1-9	4 (2.6)	8/4
Disease control	22-66	44.9 (16.2)	1-5	2 (1.6)	7/5
Normal control	22-70	43.1 (16)	—	—	6/6

- 1 Kaji R, Shibasaki H, Kimura J. Writer's cramp: a disorder of motor subroutine? *Ann Neurol* 1995;38:837-8.
- 2 Bindman E, Tibbetts RW. Writer's cramp: a rational approach to treatment? *Br J Psychiatry* 1977;131:143-8.
- 3 Kaji R, Rothwell JC, Katayama M, *et al.* Tonic vibration reflex and muscle afferent block in writer's cramp. *Ann Neurol* 1995;38:155-62.

- 4 Saxena S, Brody AL, Schwartz JM, *et al.* Neuroimaging and frontal-subcortical circuitry in obsessive-compulsive disorder. *Br J Psychiatry* 1998;35(suppl):26-37.
- 5 Broocks A, Thiel A, Angerstein D, *et al.* Higher prevalence of obsessive-compulsive symptoms in patients with blepharospasm than in patients with hemifacial spasm. *Am J Psychiatry* 1998; 155:555-7.

Cervical spondylotic myelopathy and Kennedy syndrome mimicking amyotrophic lateral sclerosis

A 62 year old male patient presented with a longstanding history of slowly progressive limb weakness, speech, and swallowing difficulties. In 1983 a diagnosis of amyotrophic lateral sclerosis had been made. At that time his physical examination showed tongue atrophy with fibrillations, proximal limb weakness, and brisk lower limb tendon reflexes. Electromyography showed abnormal "spontaneous activity" with fibrillations and positive sharp waves in muscles of all limbs. His further medical and family history was unremarkable.

In January 2000 neurological examination showed mild facial weakness, marked atrophy and fibrillations of the tongue, severe dysarthria and dysphagia, atrophy, and weakness of the shoulder girdle and arm muscles, and an unsteady and broad based gait. Apart from brisk knee jerks, deep tendon reflexes were absent and plantar responses were negative. Sensory testing was normal. General physical examination showed slight gynecomastia. Laboratory testing showed raised creatine kinase (305 U/l) and lactate dehydrogenase (195 U/l) concentrations. Needle EMG demonstrated positive sharp waves and fibrillation potentials and long duration polyphasic motor unit potentials with increased amplitudes in muscles of all limbs. By contrast, motor and sensory nerve conduction studies gave normal results.

As cervical myelopathy is an important differential diagnosis in patients with suspected motor neuron disease, cervical MRI was performed. As shown in figure 1, MRI disclosed marked cervical spondylosis with appreciable narrowing of the spinal canal between C3 and C6. In addition, T2 weighted images showed intramedullary changes with foci of high signal intensity at the level of C5 indicating myelopathy. Although these changes may readily explain the weakness in his upper limbs, the cause of bulbar symptoms and denervation in his lower limbs remained unclear.

The presence of slight bilateral gynecomastia prompted us to look for androgen receptor gene mutations, which cause X linked spinal bulbar muscular atrophy.¹ This disorder, also known as Kennedy syndrome, is caused by an unstable expansion of a CAG repeat in exon 1 of the androgen receptor gene (Xq11-12). The androgen receptor is highly expressed in motor neurons of the brain stem and spinal cord. The CAG repeat expansion is thought to confer a toxic gain of function to the androgen receptor protein resulting in irreversible damage of brain stem and spinal cord motor neurons. In addition, the impaired ability to transactivate androgen sensitive genes of the mutated receptor may account for endocrine features such as gynecomastia or testicular atrophy in spinal bulbar muscular atrophy.²⁻⁴

Genetic analysis in our patient showed one allele carrying an abnormally expanded CAG



Figure 1 Spondylotic ridging at the C3-C6 level causing spinal stenosis is shown in the sagittal T2 weighted MR image. Additionally increased signal intensity is seen within the spinal cord at the level of C5.

repeat (44; normal range 16-33) thus confirming the diagnosis of Kennedy syndrome.

The present patient with coexisting cervical spondylotic myelopathy and Kennedy syndrome highlights the diagnostic value of an intensified investigation including cervical MRI and androgen receptor gene analysis in patients with an unusual clinical presentation of motor neuron disease.

D FISCHER
U WÜLLNER
T KLOCKGETHER
R SCHRÖDER

Neurologische Klinik und Poliklinik, Rheinische
Friedrich-Wilhelms-Universität,
Sigmund-Freud-Strasse 25, 53105 Bonn, Germany

K WILHELM
Radiologische Klinik, Rheinische
Friedrich-Wilhelms-Universität, Sigmund-Freud-Str.
25, 53105 Bonn, Germany

Correspondence to: Dr D Fischer
myologie@meb.uni-bonn.de

- 1 La Spada A, Wilson EM, Lubahn DB, *et al.* Androgen receptor gene mutations in X-linked spinal and bulbar muscular atrophy. *Nature* 1991;352:77-9.
- 2 Kennedy WR, Alter M, Sung JH. Progressive proximal spinal and bulbar muscular atrophy of late onset: a sex-linked recessive trait. *Neurology* 1968;18:671-80.
- 3 Sobue G, Hashizume Y, Mukai E, *et al.* X-linked recessive bulbospinal neuronopathy: a clinicopathological study. *Brain* 1989;112:209-32.
- 4 Brooks BP, Fischbeck KH. Spinal and bulbar muscular atrophy: a trinucleotide-repeat expansion neurodegenerative disease. *TINS* 1995;18:459-61.

Posturally evoked vomiting without nystagmus in a patient with Arnold-Chiari malformation

Arnold-Chiari malformation type I (ACM I) is a developmental anomaly of the rhombencephalon characterised by displacement of the cerebellar tonsils into the foramen magnum and elongation of the medulla. It usually presents in adult life with head

motion induced oscillopsia, ataxia, headaches, cervical pain, or Valsalva induced dizziness.¹

Various ocular motor abnormalities have been reported in patients with ACM I. Among these, downbeat nystagmus and periodic alternating nystagmus are the most common. Other often encountered ocular signs, such as gaze evoked nystagmus, rebound nystagmus, and impaired smooth pursuit, reflect cerebellar involvement.^{2,3}

Vertigo of vestibular origin, being peripheral or central, is usually accompanied by nystagmus and nausea, or vomiting, and is often influenced by head position.^{3,4} The entity of central positioning vomiting without, or little, vertigo and nystagmus (posturally evoked vomiting (PEV)) was first reported by Drachman *et al* and later recognised by Brandt and Baloh and Halmagyi.^{3,5}

Posturally evoked vomiting is generally poorly known as a warning symptom of a posterior fossa lesion and is often misinterpreted.⁵ Whereas it has been documented in patients with posterior fossa tumours, it has not been reported in patients with developmental abnormalities. We report on a patient with ACM I where PEV was the most prominent presenting symptom.

This 57 year old woman was seen in our vertigo clinic because of gait unsteadiness and postural vomiting. Her history included an aortic valve replacement for aortic insufficiency, nephrolithiasis, and peptic disease. She was treated with warfarin.

For years, she had dizziness and severe nausea while looking up. During the past months severe nausea and vomiting appeared when she tilted her head to either side and down. Lately, she had become unsteady. She also complained about left high pitched tinnitus and intermittent pain in the left shoulder.

On examination her eyes were properly aligned with a full range of movements. No primary or gaze evoked nystagmus was seen, with and without Frenzel's glasses. The saccadic eye performance was normal, but the smooth pursuit in both the horizontal and the vertical plane was saccadic. The vestibulo-ocular reflex, examined by a doll's eyes movement, head thrust test, and dynamic visual acuity test, was normal. Mild dysmetria on finger-nose testing and finger-finger testing was found bilaterally. The deep tendon reflexes were brisk in the upper and normal in the lower limbs. The plantar toe responses were flexor. Sensation was normal. The gait was atactic and the Romberg test negative.

On testing the eyes in the Dix-Hallpike position to either ear, as well as in the head down position, the patient reported severe nausea, and became pale and perspired. However, no nystagmus was seen either by direct observation, or with Frenzel's glasses.

The symptoms persisted with repetition of the positioning.

An electronystagmogram (ENG) documented saccadic eye tracking in the horizontal plane. The optokinetic nystagmus was asymmetric with little increment after increased speed velocity of the target. When supine and with her head turned to the left, nystagmus of 7°/s, beating to the left, was recorded. No nystagmus was recorded on Hallpike testing. The caloric test was within normal limits.

A behavioural audiogram showed mild bilateral sensorineural hearing loss in the frequencies of 2000 Hz-8000 Hz with normal speech discrimination, interpreted as presbycusis. The brain stem auditory evoked potentials were normal.

Brain T1 and T2 weighted MRI with gadolinium enhancement disclosed low lying cerebellar tonsils with elongation of the medulla and pons, compatible with ACM I. A brain stem or cervical syrinx was not demonstrable (fig 1).

In view of the progressive symptomatology, posterior fossa decompression was considered, but postponed because of the cardiac situation of the patient. Clonazepam was offered to the patient for alleviation of the posturally evoked symptoms, but was refused because of possible sedation.

Central paroxysmal positioning vertigo occurs transiently on changing the head position, as opposed to central positional nystagmus, which persists as long as the head position is sustained.^{3,4} Central paroxysmal positioning vertigo can be differentiated from benign paroxysmal positioning vertigo by its shorter latency, longer duration of the attack, and a direction changing nystagmus which is not attributable to stimulation of a single canal.^{3,4} It usually indicates a lesion around the fourth ventricle, dorsal vermis, or diffuse cerebellar pathology.³ Central positional nystagmus is often bilateral, of low constant frequency, and occurs in association with lower brain stem or vestibulocerebellar lesions.³

In 1977 Drachman *et al* described the entity of PEV.⁵ In their two patients (one with a metastasis and the other with a glioblastoma of the cerebellar vermis) PEV was a prominent clinical sign, whereas vertigo and nystagmus diminished with the progression of the pathological process. Positional vomiting became so severe that the patients had to hold their head in a forced position to prevent vomiting. The symptoms cleared after treatment with vestibular suppressants. Drachman *et al* explained the condition of PEV by dissociation of the vomiting centres from oculomotor and receptive pathways of the complex vestibular syndrome by a brain stem infiltrating lesion.

Arbusow *et al* described a patient with amiodarone induced positional vomiting with mild vertigo, downbeat nystagmus, and limb ataxia, responsive to benzodiazepines. According to their concept, it is in keeping with that of Drachman *et al*, amiodarone disinhibits selectively the connections between the dorsal cerebellar vermis and the vomiting centres in the postremal area and the lateral

reticular formation, sparing relatively the vermis-ocular motor circuitry.⁶

Severe positioning nausea and vomiting with inconsistent dizziness and without nystagmus, was present in this patient with ACM I. Initially, nausea occurred only on head extension, but later on all head movements but anteflexion. An ENG showed only low speed positional—that is, sustained nystagmus in one head direction. On Hallpike positioning testing nystagmus was not recorded. Other ocular motor findings (saccadic pursuit and abnormal optokinetic nystagmus) were compatible with a cerebellar lesion.^{3,4}

In ACM, a normal vestibular input after changes in head position might be misprocessed by transient brain stem compression, leading to dissociation of the vomiting centres from other parts of the vestibular reflex. The relatively mild signs of positional vomiting in our patient might be due to the benign nature of the underlying disease, as opposed to the invasiveness of tumours or acute neurotoxicity, leading to severe PEV.^{5,6}

This patient with ACM seems to be unique in the literature, as PEV has not been reported in this clinical setting.

Further neurophysiological studies are needed to elucidate the pathogenesis of PEV.

L POLLAK

C KLEIN

J M RABEY

Department of Neurology, Assaf Harofeh Medical Centre, Affiliated to the Sackler School of Medicine, Tel-Aviv University, Israel

E REICHENTHAL

Department of Neurosurgery, Ben Gurion University, Beer Sheva, Israel

Correspondence to: Dr L Pollak, Neve-Nir 1, Nes-Ziona 74042, Israel
eitan.pollak@telrad.co.il

- 1 Stevens JM, Serva WAD, Kendall BE, *et al*. Chiari malformation in adults: relation of morphological aspects to clinical features and operative outcome. *J Neurol Neurosurg Psychiatry* 1993;56:1072-7.
- 2 Bronstein AM, Miller DH, Rudge P, *et al*. Down beating nystagmus: magnetic resonance imaging and neuro-otological findings. *J Neurol Sci* 1987;81:173-84.
- 3 Brandt T. *Vertigo. Its multisensory syndromes*, 2nd ed. London: Springer-Verlag 1999:199-213, 291-8.
- 4 Baloh RW, Halmaghi GM. *Disorders of the vestibular system*. New York: Oxford University Press, 1996:446-53, 488-93.
- 5 Drachman DA, Diamond ER, Hart CW. Posturally evoked vomiting: association with posterior fossa lesions. *Ann Otol Rhinol Laryngol* 1977;86:97-101.
- 6 Arbusow V, Strupp M, Brandt T. Amiodarone induced severe prolonged head positional vertigo and vomiting. *Neurology* 1998;51:917.

Lunchtime headache

Chronic primary unilateral headaches fall into one of five categories: chronic cluster headache, chronic paroxysmal hemicrania, hemicrania continua, cervicogenic headache, and SUNCT syndrome. Overlap between types is recognised. Although the differentiation of these is sometimes difficult, there are important therapeutic implications—for example, indomethacin has a dramatic effect on chronic paroxysmal hemicrania but is less effective in chronic cluster headache. Here, a patient with paroxysmal unilateral headaches, occurring precisely on the same day of the week and at the same time, is described.

A 57 year old man presented with episodic, right sided, moderate to severe headaches of 9 months' duration. He developed these always on a Monday at 1300 hours. The

headaches were sharp and throbbing with the maximum pain behind the right eye. The headache then radiated to the back of the head without crossing the midline. The pain was felt in paroxysms each lasting several minutes. He did not experience any visual symptoms, nausea, nasal congestion, lacrimation, or ptosis. Initially the total duration of the headaches was about 10 hours but it became progressively longer, remaining until Wednesday on some occasions. They were not related to the consumption of food and he did not go out drinking on Sunday nights.

At the age of 20, he had had four episodes of severe headache on the right on four consecutive days after breaking rest during a week of night shifts as a warehouseman. In between the Monday "lunchtime headaches", he remained well. He had no personal or family history of migraine. He was taking propranolol for mild hypertension. His examination was normal. There was mild cervical spondylosis on plain radiology. Brain CT showed minor atrophy.

A diagnosis of chronic paroxysmal hemicrania was made, and he was prescribed indomethacin (50 mg three times on Mondays). With this his headaches were delayed until Tuesday morning and the duration of the headaches was reduced to less than 24 hours. The headaches also became less predictable. A more protracted course of indomethacin rendered him headache free.

Review of this man's headache history suggests that the episode he had when aged 20 was probably an isolated "cluster" headache. The more recent, highly predictable, right sided headaches on Monday afternoons were unusual. The predictability of these was such that he could time it precisely to 1300 hours. Their characteristics were not classic for chronic paroxysmal hemicrania, in which attacks last between 20 and 30 minutes and are accompanied by ipsilateral nasal congestion and lacrimation.¹ Attacks of chronic paroxysmal hemicrania can, however, have clockwork regularity and are abolished by indomethacin.¹ Headaches in this case are also different from chronic cluster headache as the paroxysms occurred many times throughout the day (repetition rate 12-16/day) and the total duration of an "attack" could be up to 72 hours. Recently another variant of chronic paroxysmal hemicrania—hemicrania continua—has been described² and these headaches are non-paroxysmal and continuous. The degree of cervical spondylosis in this patient was mild, and furthermore the nature of the attacks was not that of cervicogenic headache. SUNCT syndrome is a type of paroxysmal unilateral headache consisting of short lasting, unilateral, neuralgiform headaches with conjunctival injection and tearing.^{3,4} Patients have up to 30 paroxysms/hour, occurring once or twice a day.³ This diagnosis is also highly unlikely in our patient, with his clinical presentation and therapeutic response. It was thought that this man's headaches were most probably due to chronic paroxysmal hemicrania evolving from a possible early "cluster" attack, supporting the suggestion that a common pathophysiology underlies these "trigeminal-autonomic cephalalgias".⁴ The precise predictability of the Monday afternoon headaches is fascinating, and unexplained.

B SENANAYAKE

R DE SILVA

Essex Centre for Neurology and Neurosurgery, Oldchurch Hospital, Romford, Essex RM7 0BE, UK

Correspondence to: Dr R de Silva

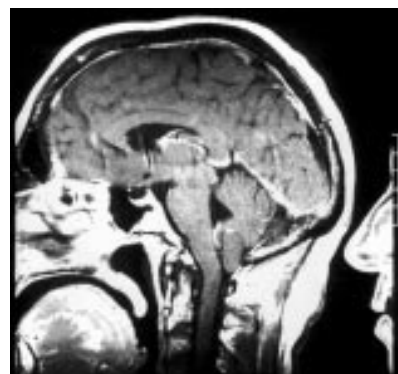


Figure 1 Brain MRI shows low lying cerebellar tonsils and elongation of the pons and medulla, compatible with ACM I.

- 1 Sjaastad O, Dale I. A new (?) clinical headache entity "chronic paroxysmal hemicrania". *Acta Neurol Scand* 1976;54:140-59.
- 2 Sjaastad O, Spierings ELH. Another headache absolutely responsive to indomethacin. *Cephalalgia* 1984;4:65-70.
- 3 Pareja JA, Sjaastad O. SUNCT syndrome, a clinical review. *Headache* 1997;37:195-202.
- 4 Goadsby PJ, Lipton RB. A review of paroxysmal hemicranias, SUNCT syndrome and other short lasting headaches with autonomic features including new cases. *Brain* 1997;120:193-209.

CORRESPONDENCE

Painful stimulation

Once again, the meaningless term "response to pain" has been allowed to appear in *the Journal*.¹

In this particular instance, the patient was unconscious and the brain stem was separated from higher centres. The stimulus may have been *noxious*, but there is no evidence whatsoever that it was painful. Furthermore, as it is insisted, rightly, that other sensory stimuli be fully and accurately described, why is "pain" permitted without qualification? This often means pricking with a pin. Unless the pin or needle is driven in and twisted, the modality tested is in fact sharpness discrimination and not pain—the two are dissociated in some lesions. Mechanical noxious stimulation, which is interpreted as painful in the intact and fully conscious person, may be brought about by skinfold pinch or various other manoeuvres; thermal and chemical noxious stimulation also, of course, exist. But unless the nature of the stimulus is specified, the expression *noxious stimulation* (let *pain* and *painful stimulation* be forever abolished) is not only almost devoid of relevance, but misleading.

It also seems unfortunate that the recent *Journal of Neurology, Neurosurgery, and Psychiatry/Association of British Neurologists* supplement on stroke did not mention central poststroke pain. This difficult neurological sequel occurs in some 8%–10% of survivors of stroke^{2,3} (6000–7000 new cases/year in the United Kingdom). Its correct diagnosis, treatment, and possible prevention should be a matter of great concern to neurologists.

D BOWSHER

Pain Research Institute, University Hospital Aintree,
Lower Lane, Liverpool L9 7AL, UK

Correspondence to: pri@liv.ac.uk

- 1 Wijdicks EFM, Atkinson JLD, Okazaki H. Isolated medulla oblongata function after severe traumatic brain injury. *J Neurol Neurosurg Psychiatry* 2001;70:127-9.
- 2 Andersen G, Vestergaard K, Ingeman-Nielsen M, et al. Incidence of central post-stroke pain. *Pain* 1995;61:187-94.
- 3 Bowsher D. Stroke and central post-stroke pain in an elderly population. *Journal of Pain* 2001 (in press).

Is it really Alzheimer's disease?

We read the recent article of Cappa *et al* with great interest.¹ Using HMPAO SPECT, the authors studied 24 patients diagnosed as having (probable) Alzheimer's disease (AD) on the basis of DSM-III-R and the

NINCDS-ADRDA criteria. According to Butters *et al*, they distinguished, on the basis of neuropsychological tests, patients with a diffuse pattern of cognitive deficits (dAD) from those with focal temporal lobe dysfunction (FTLD).² Patients with FTLD are reported by both studies as having a better cognitive prognosis. Whereas the dAD group had diffuse perfusion deficits, the patients with FTLD showed a circumscribed reduction of tracer uptake in the left or right, or both temporal lobes.

We comment on the problem of suggesting AD in these patients with dementia. In the study of Butters *et al*, seven of the patients of the FTLD group underwent necropsy. Six had the typical histopathological signs of AD. One patient, however, showed the features of hippocampal sclerosis (HS) and not those of AD (Cappa *et al* wrongly state that all necropsied cases of Butters *et al* had AD). Two major groups of patients with the histopathological signs of HS have been described in the literature: (1) HS is the most common morphological substrate of medial temporal lobe epilepsy (MTLE). The affected patients are known to show characteristic memory impairment, which correlates well with the degree of pathological changes. Progress of memory impairment in MTLE is very slow and other cognitive functions are mostly preserved.³ Most (but not all) of them have an abnormal MRI signal and HMPAO SPECT usually shows temporal hypoperfusion.⁴ (2) Patients with HS have been described in necropsy series of patients with dementia.⁵ It is unknown whether these patients could have been identified by MRI, whether they had abnormal brain perfusion, and what the perfusion pattern would have been.

Cappa *et al* did not exclude patients with epilepsy, and it is not clear if their MRI procedure included high resolution T2/FLAIR sections angulated along or perpendicular to the hippocampus, which considerably increases the sensitivity for detection of MTLE-HS. They also had no histopathological evidence of AD. Thus, the authors may have included an unknown number of patients who actually did not have AD, but, for example, had temporal lobe epilepsy or dementia with the morphological substrate of HS. It is noteworthy that in a recent histopathological study on the causes of dementia (with further references)⁶ it was noted that fewer patients than clinically suspected fall into the histopathological category of AD but—among others—fall into the category of HS.

C G BIEN

C HELMSTAEDTER

C E ELGER

University of Bonn, Department of Epileptology,
Sigmund-Freud-Str. 25, 53105 Bonn, Germany

Correspondence to: Dr C G Bien
c.bien@uni-bonn.de

- 1 Cappa A, Calcagni ML, Villa G, et al. Brain perfusion abnormalities in Alzheimer's disease: comparison between patients with focal temporal lobe dysfunction and patients with diffuse cognitive impairment. *J Neurol Neurosurg Psychiatry* 2001;70:22-7.
- 2 Butters MA, Lopez OL, Becker JT. Focal temporal lobe dysfunction in probable Alzheimer's disease predicts a slow rate of cognitive decline. *Neurology* 1996;46:687-92.
- 3 Helmstaedter C, Elger CE. The phantom of progressive dementia in epilepsy. *Lancet* 1999;354:2133-4.
- 4 Menzel C, Hufnagel A, Grunwald F, et al. The relevance of interictal rCBF brain SPECT in temporal lobe epilepsy: diagnostic value and

effects of spatial resolution. *Ann Nucl Med* 1995;9:215-23.

- 5 Corey-Bloom J, Sabbagh MN, Bondi MW, et al. Hippocampal sclerosis contributes to dementia in the elderly. *Neurology* 1997;48:154-60.
- 6 Crystal HA, Dickson D, Davies P, et al. The relative frequency of "dementia of unknown etiology" increases with age and is nearly 50% in nonagenarians. *Arch Neurol* 2001;57:713-19.

Cappa, *et al* reply:

We are grateful to Bien, Helmstaedter, and Elger as they allow us to clarify a point of our study that was not explicitly discussed in the paper recently published in this *Journal*.¹

We obviously agree with them that, in the absence of histopathological confirmation, the diagnosis of "probable" Alzheimer's disease (AD) can be questioned on methodological grounds, particularly in patients with focal temporal lobe dysfunction (FTLD).

However, we think it rather implausible that some of our patients with FTLD were in reality affected by other non-AD diseases, in particular by those suggested by Bien *et al*: medial temporal lobe epilepsy (MTLE) or dementia sustained by hippocampal sclerosis (HS).

As for MTLE, which Bien *et al* consider a relevant source of possible errors in the diagnosis of patients with AD and FTLD,² we emphasise that none of our patients—with diffuse cognitive impairment (dAD) or with FTLD—was affected by epilepsy. In our paper, the exclusion of epileptic patients was implicit in the statement that all our patients met the NINCDS-ADRDA criteria for probable AD (according to these criteria, patients who have epilepsy could at the most attain a diagnosis of possible AD).

Apart from MTLE, we are certainly aware that HS is sometimes responsible for a dementia syndrome mimicking AD, as recently discussed in several neuropathological studies.³⁻⁶

On the other hand, a careful review of these studies leads to the following conclusions: (1) in a general demented population, pure HS (HS without Alzheimer-type or other well characterised degenerative or vascular pathological changes) seems to be a very rare cause of dementia. Ala *et al*³ found only 0.4% of pure HS in a retrospective study of 1771 unselected demented patients and Jellinger⁷ obtained similar results (0.53%) in a consecutive necropsy series of 746 demented subjects older than 55 years! (2) HS significantly contributes to dementia only in old or very old (≥ 80 years of age) demented patients with documented cardiovascular diseases (for example, ischaemic heart disease, arrhythmias, congestive heart failure in about 88% of cases) or with depression as another frequent (63%) clinical feature.^{4,6} (3) When HS is associated with other neuropathological changes, the precise boundaries with other forms of dementia are often less clear and the differential diagnosis may be controversial even at a pathological level. In the necropsy series of Corey-Bloom *et al*,⁴ for instance, more than 50% of patients with HS had enough amyloid plaques to meet NIA criteria⁸ for AD; and even using the more conservative criteria of CERAD,¹⁰ four patients out of eight could have been diagnosed as AD. (4) It is quite uncommon for demented patients with postmortem evidence of HS to have previously received a clinical diagnosis of probable AD (in most cases, patients with HS had been diagnosed as possible AD⁹).

Patients reported in our study as affected by AD with FTLT were clearly different from those described in studies claiming a significant contribution of HS to dementia.⁴⁻⁶ Their mean age was 67.3 (SD 8.1). All of them had received a diagnosis of probable AD according to the NINCDS-ADRDA criteria. None of them had a history of cardiovascular disease or depression. At MRI—performed with high resolution T2/FLAIR sections, using transverse and coronal planes to obtain a good visualisation of the mesial temporal cortex (hippocampal and parahippocampal gyri)—all of them exhibited atrophic changes consistently involving and yet exceeding the hippocampal structures, in the absence of focal lesions attributable to vascular or other diseases. With regard to this last point, it seems to us noteworthy that Helmstaedter and Elger,² studying 63 patients with chronic pharmacoresistant MTLE sustained by HS, found significant difficulties with episodic memory tasks (learning, retrieval, and recognition of a word list), but not with semantic memory tasks (vocabulary). In our FTLT group, conversely, the presence of semantic memory impairment in association with episodic memory deficit was in agreement with the typical progression of AD pathological process, spreading from the mesial temporal areas to the adjacent temporal neocortex.¹¹ Incidentally, in four patients with FTLT who completed a clinical and neuropsychological follow up of at least 3 years an evolution towards a more diffuse pattern of cognitive impairment has now been found. In two other patients with FTLT, who recently underwent neuropsychological retesting after a 12–15 month interval from the baseline assessment, a significant worsening of cognitive deficits, which nevertheless still remained relatively circumscribed to episodic and semantic memory, has been documented.

In conclusion—and also in the light of the above cited studies investigating the relations between HS and dementia—we think that the clinical characteristics of our patients with FTLT were hardly explainable in terms of an underlying HS and more consistent with the diagnosis of probable AD.

G VILLA
A CAPPA
C MARRA
G GAINOTTI

*Servizio di Neuropsicologia, Istituto di Neurologia,
Università Cattolica del Sacro Cuore, Policlinico A
Gemelli, Largo Agostino Gemelli 8, I-00168 Roma,
Italy*

Correspondence to: Dr G Villa
iclnp@unicatt.rm.it

- 1 Cappa A, Calcagni ML, Villa G, *et al.* Brain perfusion abnormalities in Alzheimer's disease: comparison between patients with focal temporal lobe dysfunction and patients with diffuse cognitive impairment. *J Neurol Neurosurg Psychiatry* 2001;70:22–7.
- 2 Helmstaedter C, Elger CE. The phantom of progressive dementia in epilepsy. *Lancet* 1999; 354:1225–30.
- 3 Ala TA, Beh GO, Frey WH 2nd. Pure hippocampal sclerosis: a rare cause of dementia mimicking Alzheimer's disease. *Neurology* 2000;54:843–8.
- 4 Corey-Bloom J, Sabbagh MN, Bondi MW, *et al.* Hippocampal sclerosis contributes to dementia in the elderly. *Neurology* 1997;48:154–60.
- 5 Crystal HA, Dickinson D, Davies P, *et al.* The relative frequency of "dementia of unknown etiology" increases with age and is nearly 50% in nonagenarians. *Arch Neurol* 2000;57:713–7.
- 6 Dickinson DW, Davies P, Bevana C, *et al.* Hippocampal sclerosis: a common pathological feature of dementia in very old (> or = 80 years of age) humans. *Acta Neuropathol (Berl)* 1994; 88:212–21.

- 7 Jellinger K. Hippocampal sclerosis: a common pathological feature of dementia in very old humans. *Acta Neuropathol* 1994;88:599.
- 8 Jellinger K. Pure hippocampal sclerosis: a rare cause of dementia mimicking Alzheimer's disease. *Neurology* 2000;55:739.
- 9 Hyman BT, Trojanowski JQ. Consensus recommendations for the postmortem diagnosis of Alzheimer disease from the National Institute on Aging and the Reagan Institute Working Group on diagnostic criteria for the neuropathological assessment of Alzheimer disease. *J Neuropathol Exp Neurol* 1997;56:1095–7.
- 10 Gearing M, Mirra S, Hedren J, *et al.* The consortium to establish a registry for Alzheimer's disease (CERAD). Part X. Neuropathology confirmation of the clinical diagnosis of Alzheimer's disease. *Neurology* 1995; 45:461–6.
- 11 Hodges JR, Patterson K. Is semantic memory consistently impaired early in the course of Alzheimer's disease? Neuroanatomical and diagnostic implications. *Neuropsychologia* 1995; 4:441–59.

BOOK REVIEWS

Intractable focal epilepsy. Edited by JOHN OXBURY, CHARLES POLKEY, and MICHAEL DUCHOWNY (Pp 878, £125.00). Published by WB Saunders, London, 2000. ISBN 0-7020-2428-7.

This is quite a comprehensive multiple author textbook which sets out in detail clinical, pathological, investigative, and medical and surgical management of patients with focal epilepsy. The standpoint of the editors is to use the words "focal epilepsy" rather than partial epilepsy because they think that it is more descriptive of the probable underlying pathophysiological changes. As such, the book is of particular interest to epileptologists and other specialists likely to be involved in developing an epilepsy surgery programme. Many of the other epilepsies would of course have more widespread or multifocal underlying substrates or generator circuits.

The selection of the editors for individual chapters incorporates a diverse range of specialists principally from Europe and North America. As such there is a useful diversity of views represented and many of the authors selected are acknowledged world experts in the areas of their chapter.

The text is clearly written with good illustrations and high quality photographs. There is a standard useful summary of key points with each chapter, and clearly there has been significant editorial input to produce an easily readable textbook with common themes.

The text covers many of the areas needed to support an epilepsy surgery programme, although it might be advantageous in a future edition to include further information about the rapidly expanding field of neurogenetics and epilepsy. The overall quality of the material would more than justify its inclusion in a specialist library or to support physicians or associated specialists working in the field of epilepsy surgery.

DAVID FISH

Schizophrenia in children and adolescents. Edited by HELMUT REMSCHMIDT (Pp 924, £39.95). Published by Cambridge University Press, Cambridge, 2000. ISBN 0 521 79428 5.

There are few psychiatric conditions as devastating as childhood schizophrenia. Case descriptions became available soon after schizophrenia was first recognised as an entity but a childhood onset is fortunately rare. Early samples probably included children with autistic spectrum disorders as these were only recognised in the 1940s. In spite of shared deficits there are important clinical differences between autism and childhood schizophrenia. The wealth of research into autism has not been matched in the field of childhood schizophrenia, perhaps because of its rarity but also because clinicians can draw on the findings on adult disorders. This book is written by experts in the field and helpfully describes current thinking and knowledge in childhood schizophrenia. It includes a chapter on historical aspects, and specially useful for clinicians, developmentally informed guidelines on differential diagnosis and management. There is a full discussion on the use of traditional and newer anti-psychotic medication.

Authors make well the case for the similarities between childhood and adult schizophrenia, they document the empirical evidence indicating increased pre-morbid developmental and personality anomalies, perhaps a stronger genetic vulnerability and worse outcome with a childhood onset. Research findings are detailed on information processing deficits and on anomalies of thought processes comparable to those seen in adult patients. The important on-going research by the National Institute of Mental Health is referenced. Outstanding areas for future research are identified. This should attempt a better description of pre-morbid developmental and behavioural anomalies currently subsumed under a bewildering array of overlapping syndromes and their differentiation from those seen in autism.

This book represents a welcome update on research and clinical thinking in childhood schizophrenia and will be useful to clinicians.

ELENA GARRALDA

Charcot-Marie-Tooth disease. A practical guide. Compiled by CMT INTERNATIONAL UK (Pp 113, £10.00). Published by CMT International UK, Penarth, 2000. ISBN 0 9533883 0 1.

Charcot-Marie-Tooth disease. A practical guide, is a book compiled by CMT International UK with the aim of providing an overview of Charcot-Marie-Tooth disease (CMT) with a particular emphasis on providing practical day to day advice for living with the disease. It is aimed at doctors and patients and other people involved with CMT. It is well written and excellently presented and provides a range of information that the intended audience will find invaluable.

The book is divided into three main sections. The first section deals with genetic and medical issues. The known genetic variants of the disease are well described and accurate except for one mistake stating that the gene duplication that causes CMT1a is on chromosome 22 when it is actually on

chromosome 17. I thought the section covering CMT inheritance was particularly well presented and illustrated using simple diagrams to explain the various inheritance mechanisms. There was also a very useful glossary of scientific and medical terms in this section.

The second section deals with living with CMT. This covers many important areas for the patient including coming to terms with the diagnosis, care of the feet, pain control, and secondary complications. Foot deformities and their surgical correction were particularly well covered.

The third section deals with practical issues including finding work, having a baby, driving, and CMT and aids to daily living. This section is particularly useful in providing contact details for many different organisations who will help patients. All three sections are supported by informative appendices.

This book is an excellent patient oriented guide, full of useful information and contacts. It will be a particularly useful book to recommend to newly diagnosed patients.

MARY REILLY

Epilepsy in clinical practice. A case study approach. Edited by ANDREW N WILNER (Pp272, US\$34.95). Published by Demos Medical Publishing, New York, 2000. ISBN 1-888799-34-x.

This is an interesting book, directed at a wide audience including neurologists, medical students, social workers, and pharmacists, which sets out to demonstrate the management of epilepsy in practice by presenting case histories of people with seizures. The issues addressed include such common clinical difficulties as diagnosis (and the problem of non-epileptic attacks), epilepsy in pregnancy, withdrawal of antiepileptic drugs, and status epilepticus. Alternative therapies and psychiatric issues are also discussed. Each history is followed by questions about the management of the patient, which the author answers according to his practice, with appropriate explanations. The second part of the book consists of a sizeable *Resources* section providing such diverse information as the classifications of epileptic seizures and syndromes, seizure history checklist, home safety checklists, lists of drugs and their modes of action, websites for physicians and patients, lists of epilepsy centres, and driving regulations within the United States.

Both sections of the book contain a wealth of information, and the histories, which are easy to read, provide a useful insight into the problem areas of epilepsy, together with possible solutions. Inevitably the views given describe the author's personal practice, and the nature of the book does not allow detailed examination of the evidence underlying the decision making. However, it emphasises the importance of tailoring treatment to the individual patient, and addresses the social issues in a manner often missing from larger texts. I found the fact that the book was clearly directed at an American audience, which was apparent not only in the *Resources* section, but also colouring the choice of medication and

the advice given (for example, on driving), somewhat distracting. Nevertheless, it should prove useful reading for those involved in the care of people with epilepsy.

YVONNE HART

Clinical neuroanatomy for medical students, 5th edition. By RICHARD S SNELL (Pp 545, £29.95). Published by Lippincott Williams and Wilkins, Baltimore, 2001. ISBN 0 7817 2831 2.

Richard Snell's *Clinical anatomy for medical students* is a successful clinically oriented text that contains essential facts and explanations without excessive detail. His parallel neuro-anatomical text has similar aims.

Each chapter begins with chapter objectives; these are actually a brief explanation of broad aims for learning rather than specific educational objectives. There follow the main anatomical information of the chapter. New terms are printed in bold text, a style that is less than effective as attention is drawn to these rather than to key points of understanding. Next comes an extensive section of clinical notes that constitute the greatest strength of the book. They make clear the ways in which anatomical knowledge underpins diagnosis and management, and provide the student with valuable motivation to gain control of the key facts. Understanding is then tested by a set of clinical problem solving scenarios with answers. The last element is a set of review questions in a multiple choice format, many of which test topographical neuro-anatomy at an excessively detailed level.

The first three chapters cover gross anatomy and cell biology of the nervous system and the fundamentals of nervous system function. These take things fairly gently and a good deal of the material will be familiar to a medical student who already has a reasonable knowledge of anatomy and physiology. The material in the functional chapter in particular is rather variable in level and quality.

The next four chapters provide a fairly conventional ascending regional treatment of the CNS from spinal cord through brain stem and cerebellum to cerebrum. The author's intention to provide an essential core of knowledge has here been overwhelmed by reluctance to leave anything out.

The next nine chapters are much better. These deal with the nervous system in a systems based way, covering such themes as cortical organisation, the reticular formation, the limbic system, and the hypothalamus, with balance and clarity. The chapter on the autonomic system will help students who find the principles difficult, although it is disappointing that the afferent component of the parasympathetic system is not considered as an entity. The coverage of cranial nerves suffers from insufficient explanation of the components of a cranial nerve—the term “branchiomotor” is used but not explained. The final chapter, on development of the nervous system, seems to have been included for completeness rather than out of any conviction that it matters.

Illustrations are very variable in quality, with excellent three dimensional half tone

drawings and good illustrations of pathways but some poor thumbnail sketches. Some illustrations have been recycled from the author's anatomy text without adaptation for their new context. Imaging material is varied and usually well chosen. The tables tend to be congested with information and are not likely to be found helpful.

The short time available for studying neuroanatomy in most curricula means that students tend to use textbooks to clarify the deficiencies of their lecture notes rather than for systematic reading. The index is fairly comprehensive and will usually take the reader to the right place.

In summary, Professor Snell has not yet produced the compact, clear guide to the anatomical basis of clinical neurology needed by modern medical students; the message is still buried in excessive anatomical detail and terminology. However, his book deserves recommendation for its clinical notes. These could provide the starting point for defining the core anatomical knowledge in a future edition.

Clinical neuroanatomy. An illustrated review with questions and explanations, 3rd edition. By RICHARD S SNELL (Pp 256, £19.95). Published by Lippincott Williams and Wilkins, Baltimore, 2001. ISBN 0 7817 2989 0.

This text recycles information and illustrations from his larger textbook in a form designed for rapid revision and as a reminder of forgotten neuroanatomy for clinical attachments and recent graduates. Use of clinical notes and review questions parallels the larger text, though the chapter organisation is somewhat different. The factual content has been reduced in length, often to sequences of short statements closer to lists than to explanation. The quality of illustration is also considerably reduced by exclusion of colour.

The result is about half the length of the main text and two thirds of the price. It is hard to imagine why a student or junior doctor who has used the larger book or some other clinical anatomy text would wish to buy this considerably inferior product, still far too large to provide headlines for storage in short term memory.

ANTHONY FIRTH

CORRECTION

Frisoni GB. Structural imaging in the clinical diagnosis of Alzheimer's disease: problems and tools. *J Neurol Neurosurg Psychiatry* 2001;70:711-18. The following acknowledgments should have appeared in this editorial:

“The ideas of this paper have arisen from extensive discussion with Marco Trabucchi. I am also indebted to Alberto Beltramello, Charles DeCarli, Mikko Laakso, and Hilka Soininen for helpful suggestions and comments”